

Food Protein-Induced Enterocolitis Syndrome (FPIES)

A rare type of *non IgE mediated* food allergy affecting the gastrointestinal (GI) tract.

FPIES is a rare condition and will require a careful diagnostic evaluation, typically by an FPIES-familiar specialist, such as a Pediatric Allergist or Gastroenterologist.

Two Ways Infants/Children Might Come to Medical Attention:

Acute Presentation	Chronic Presentation
Repetitive, profound vomiting (~1-6 hrs post ingestion)	Chronic watery diarrhea with blood/mucus
Diarrhea (~2-10hrs post ingestion)	Intermittent/Chronic vomiting
Dehydration	Failure to Thrive
Lethargy	Weight loss
Pallor (pale skin, blue/gray tones)	Lethargy
Hypotension	Pallor
Abdominal Distention	Abdominal distention

^{*}note that presentation may be **acute** or **chronic** in nature and some children may have a combination both

Keep in mind that although these symptoms may be seen with an FPIES diagnosis, just as in other allergies, **there can be a very wide** range of symptoms and severity of symptoms.

Quick Facts:

FPIES is a type of **non-IgE mediated** food allergy affecting the gastrointestinal (GI) tract.

Classic symptoms of FPIES include **profound vomiting**, diarrhea, and dehydration. These symptoms can lead to severe lethargy, change in body temperature and blood pressure.

Unlike typical food allergies, **symptoms may not be immediate** and do not show up on standard allergy tests

A *clinical diagnosis*, there are no tests to confirm this diagnosis that is made by excluding other potential diagnosis and carefully considering the patient's history.

Symptoms can **mimic**: Eosinophilic Esophagitis, Eosinophilic Gastrointestinal Disorders, Fructose Malabsorption, Congenital Sucrase-Isomaltase Deficiency, Celiac Disease, Mast Cell disorders and other metabolic disorders.

Symptoms generally **first** present when a child is an infant or young toddler; some children present first with formula feeding and others with introduction of solids, reactions through breast milk may also be present.

Typically produces a **negative** allergy evaluation. It is an allergic immune reaction for which the diagnostic tests found in the allergist's office cannot be used to diagnosis FPIES and which may delay the diagnosis and take the focus off the causative food.

For additional information, please visit www.thefpiesfoundation.org



The parent may report:

"When I gave baby cereal for the first time, my child seemed to experience symptoms of a stomach bug"

"My infant takes several months to outgrow his/her clothes"

"My child spits up in very large amounts-- I worry that he/she isn't keeping much of the milk/formula down"

"My child has more frequent 'blowouts' than I have ever seen"

"My child seems very disinterested in/ seems fearful of food"

In addition, they may report other symptoms commonly noted for intolerances (rashes, reflux, inconsolable crying (day/night), frequent night waking).

Red Flags That Your Client May Be Experiencing FPIES Symptoms:

- ➤ He/she experiences episodes of severe vomiting, often occurring "out of the blue" a few hours after eating
- ➤ He/she experiences frequent diarrhea and/or blood in stools
- ➤ He/she presents as failure to thrive; although this is not always present and may be more likely with multiple triggers or chronic reactions (i.e. to milk-based formula).
- ➤ He/she has had episodes of lethargy/shock-hours after feeding that have resulted in ER visits.

FPIES Triggers, Reactions and Possible Testing:

- Some individuals affected by FPIES can react to 1-2 triggers while others may react to multiple triggers.
- > Initial reactions may present first with formula feeding while others not until introduction of solids, some infants may even experience reactions through breastmilk.
- Although *any* food can trigger a reaction, **common triggers in the US** include: rice, oats, dairy, soy.
- > Triggers can even be found in non-food items, such as craft supplies, and young children may put these items in their mouths.
- Reactions occur 2-6+ hours post-ingestion of the trigger food
- > FPIES reactions can be severe and may require emergency medical attention.
- An allergist may perform routine allergy testing on individuals undergoing an FPIES evaluation; however it is most typical for all allergy testing to produce negative result as FPIES does not show up on typical allergy diagnostic testing. This may be done to rule out IgE allergies or to monitor for the development of an IgE allergy.
- ➤ A gastroenterologist may recommend specific tests or procedures to rule out various diagnosis; procedures may include upper endoscopy, upper GI series, and sigmoidoscopy/colonoscopy.
- Atopy Patch Testing (APT) has been used as a possible guide, but studies are not clear if this is an accurate tool for FPIES.

Accessing the Existing Medical Literature:

For a comprehensive list of the latest medical journal articles on FPIES, visit: www.fpiesfoundation.org/fpies-medical-literature

For additional information, please visit www.thefpiesfoundation.org



Support for families of those affected by this condition is crucial. FPIES is often misunderstood and families may encounter community members, even medical professionals, not familiar with this diagnosis and as a result, may receive mismanaged and/or inadequate care.

Dietitian's role:

- Acknowledge support
- Ensure adequate growth and development in restricted diets.
- Suggestions of foods to try to obtain balanced diet, calorie and protein requirements,
- Micronutrient requirements on restricted diets
- Recipe modification with limited ingredients,
- Tube feeding protocols if needed,
- Monitoring of growth
- Communication with the medical team
- Give hope!

Goals to address with a family undergoing an FPIES evaluation (or newly diagnosed):

- Food Journaling
- ➤ Label reading and potential cross-contamination when selecting food products
- ➤ Alternative recipes and cooking resources
- Age-appropriate meal planning strategies for restricted diets
- Importance of nutritional diversity despite a restricted diet
- Breastfeeding support and resources
- ➤ Promote, when needed, the benefits of medical food and other supplements to improve the nutritional completeness of a patients diet
- > Encourage families, caregivers, and patients to ask for support when needed
- ➤ Help with introduction of new foods (may be done with Allergist guidance)

Additional nutrition resources:

- Food Journals & Symptom logs: http://fpiesfoundation.org/toolboxfood-journal/
- Label Reading: http://fpiesfoundation.org/cooking-and-recipes/label-reading-tutorial/
- Cooking resources: http://fpiesfoundation.org/cooking-and-recipes/
- ➤ Find resources for families: http://fpiesfoundation.org/cooking-and-recipes/nutrition-resources-for-families/

To access materials about FPIES for distribution to families and professionals, visit: www.fpiesfoundation.org/foundation-flyer or email your mailing address and written request to contact@thefpiesfoundation.org and request hard copies.



References & Articles of interest:

- 1. Kaya, A., M. Toyran, et al. (2016). "Food Protein-Induced Enterocolitis Syndrome In Two Exclusively Breastfed Infants." Pediatric Allergy and Immunology.
- 2. Mane, Shikha K., and Sami L. Bahna. "Clinical Manifestations of Food Protein-Induced Enterocolitis Syndrome." Current Opinion in Allergy and Clinical Immunology 14.3 (2014): 217–221. PMC.
- 3. Mane, S. K., M. E. Hollister, et al. (2014). "Food protein-induced enterocolitis syndrome to trivial oral mucosal contact." European Journal of Pediatrics
- 4. Monti, Giovanna et al. Food protein-induced enterocolitis syndrome by cow's milk proteins passed through breast milk. Journal of Allergy and Clinical Immunology, Volume 127, Issue 3, 679 680 (2011).
- 5. John M. Kelso, MD, and Hugh A. Sampson, MD. "Food protein-induced enterocolitis to casein hydrolysate formulas". J Allergy Clin Immunol December 1993.

Authors and Contributors:

Jen Hertsch MS, MBA

Amanda LeFew MT-BC

Bailey Koch, RD, CSP, LD

Heather Martin, RD, LD

Joy Meyer, DTR

Medical Advisory Board of The FPIES Foundation

The Medical Advisory Board of The FPIES Foundation

Allergist/Immunologist: Sakina S. Bajowala, MD J. Andrew Bird, MD Harumi Jyonouchi, MD John J. Lee, MD Fred Leickly, MD, MPH David R. Naimi, DO Luqman Seidu, MD Wayne Shreffler, MD, PhD

Gastroenterologists:
Glenn T. Furuta, MD
Benjamin D. Gold, MD
Sergio Negre Policarpo, MD, PhD
Qian Yuan, MD
Dietitians:
April Clark, RD/LD

April Clark, RD/LD Bailey Koch, RD, CSP, LD Tara McCarthy, MS, RD, LDN

To access materials about FPIES for distribution to families and professionals, visit: www.fpiesfoundation.org/foundation-flyer or email your mailing address and written request to contact@thefpiesfoundation.org and request hard copies.